Operative Techniques in
Thoracic and
Cardiovascular Surgery

Repair of Tetralogy of Fallot with Absent Pulmonary Valve Syndrome

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Tetralogy of Fallot (TOF) with absent pulmonary valve syndrome (APVS) occurs in 5% of patients with TOF. TOF-APVS includes an anterior malalignment ventricular septal defect (VSD) similar to TOF with pulmonary stenosis. The right ventricle is hypertrophied and may contain obstructing muscle bands in addition to infundibular narrowing. The pulmonary annulus is mildly to moderately hypoplastic with vestigial nubbins of nonfunctional myxomatous tissue rather than developed valve leaflets. The size of the main pulmonary artery ranges from normal to aneurysmal depending on the severity of the anomaly, whereas the branch pulmonary arteries are markedly aneurysmal, often two to three times their normal diameters. The hilar portions of the pulmonary arteries may also be dilated. The segmental branches are of normal size and appear as multiple orifices arising from the dilated portion of the artery when viewed from inside the vessel.

A patent ductus arteriosus is never present in TOF-APVS. The combination of a VSD, pulmonary insufficiency, and a patent ductus arteriosus would potentially produce physiologic aortic insufficiency: left ventricular output could flow retrograde through the ductus, into the right ventricle, across the VSD, and back into the left ventricle. It has been proposed that the absence of a pulmonary valve influences premature closure of the ductus, or the fetus would not survive. The highly pulsatile flow through the pulmonary arteries may result in the aneurysmal dilation.[1,2]

Patients with TOF-APVS can be divided into two groups. The first group presents in the neonatal period with respiratory distress, attributed to compression of the tracheobronchial tree by the aneurysmal pulmonary arteries. These patients require early surgery to avoid morbidity and death from respiratory distress and heart failure. The second group of patients is less symptomatic in infancy and may be identified later in life with symptoms of congestive heart failure, or classic TOF cyanotic spells. These patients can be managed similarly to other patients with TOF and undergo elective repair.

There are two major debated issues regarding the surgical management of neonates with TOF-APVS: management of the aneurysmal pulmonary arteries, and placement of a pulmonary valve in the right ventricular outflow tract. Multiple operations have been proposed to address these issues.[3-10] In the operation we describe below, we remove the maximal amount of abnormal pulmonary arterial tissue and place a right ventricular to pulmonary artery homograft conduit. Complete excision of the main and branch pulmonary arteries removes almost all aneurysmal tissue, thereby minimizing any chance of persistent tracheobronchial compression. Use of a homograft places a valve in the pulmonary outflow tract, which simplifies the postoperative management of pulmonary hypertension.

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Operative Technique

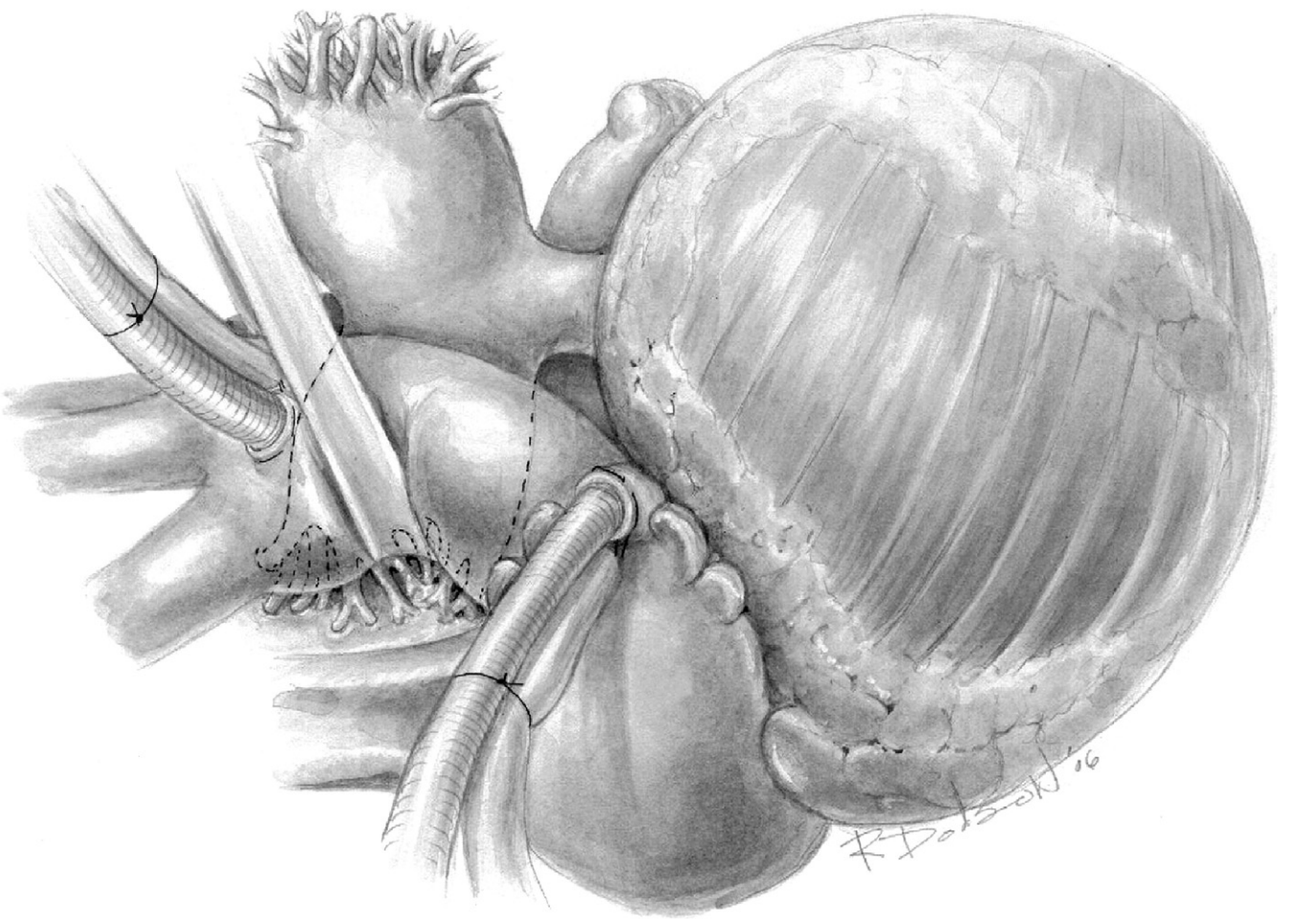

Figure 1 The chest is entered through a median sternotomy and the thymus gland is removed. The pericardium is opened and a portion is harvested and placed in glutaraldehyde for later use. Examination of the external cardiac anatomy shows the main pulmonary artery to be slightly enlarged and the right and left pulmonary arteries to be quite dilated, equal to or larger in size than the aorta. The segmental branches of these vessels are small. For larger patients we utilize aorto-bicaval cannulation and continuous cardiopulmonary bypass. For smaller patients, especially those with small superior venae cavae, we place aortic and right atrial cannulae. The patient is then cooled to 18°C. During cooling, the branch pulmonary arteries are mobilized out to the hilar branches. The aorta is then cross-clamped and 30 mL/kg of cold blood cardioplegia is administered via the aortic root. We then perform the operation under circulatory arrest with 2-minute periods of intermittent perfusion every 15 minutes. Additional doses of cardioplegia are given at these times as well.

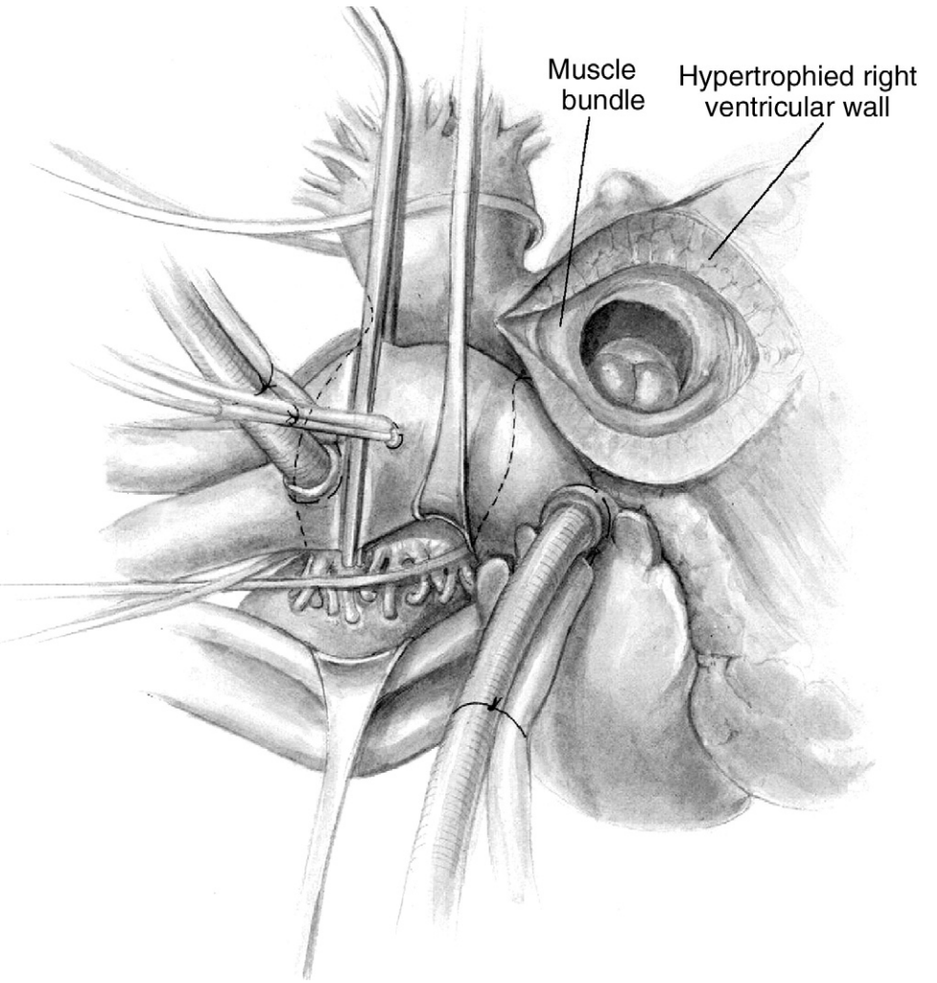

Figure 2 A longitudinal incision is made in the main pulmonary artery and continued proximally across the mildly to moderately hypoplastic pulmonary annulus and into the infundibulum of the right ventricle. This incision allows visualization of the anterior malalignment ventricular septal defect and obstructing muscle bundles in the right ventricular outflow tract. The nonfunctional pulmonary valve leaflets can be seen as well.

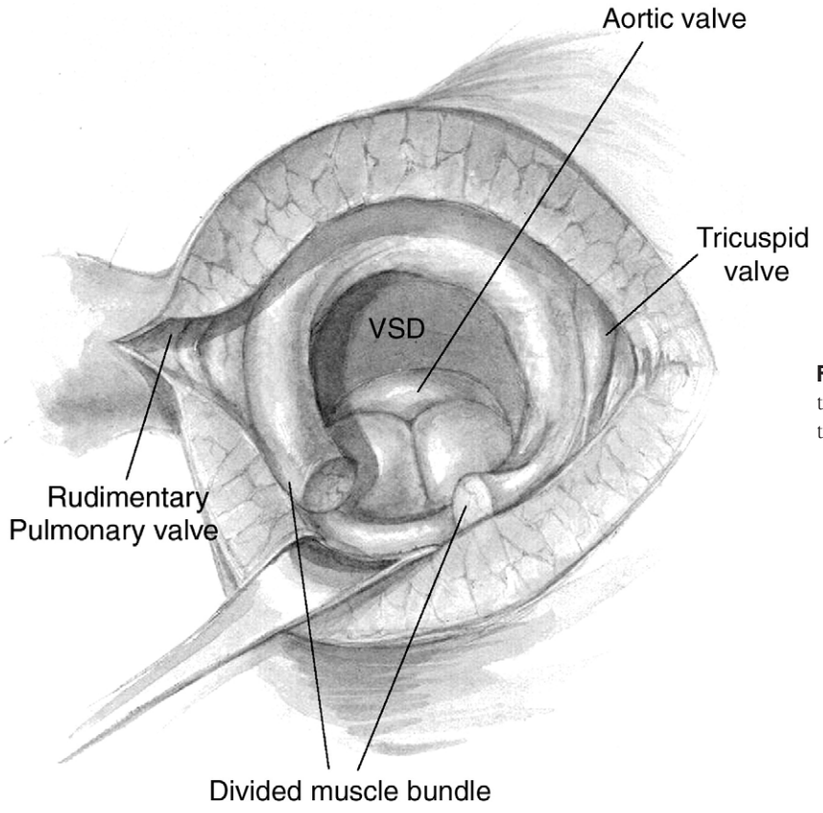

Figure 3 Infundibular muscle bundles are divided so there is no obstruction of flow out of the right ventricle.

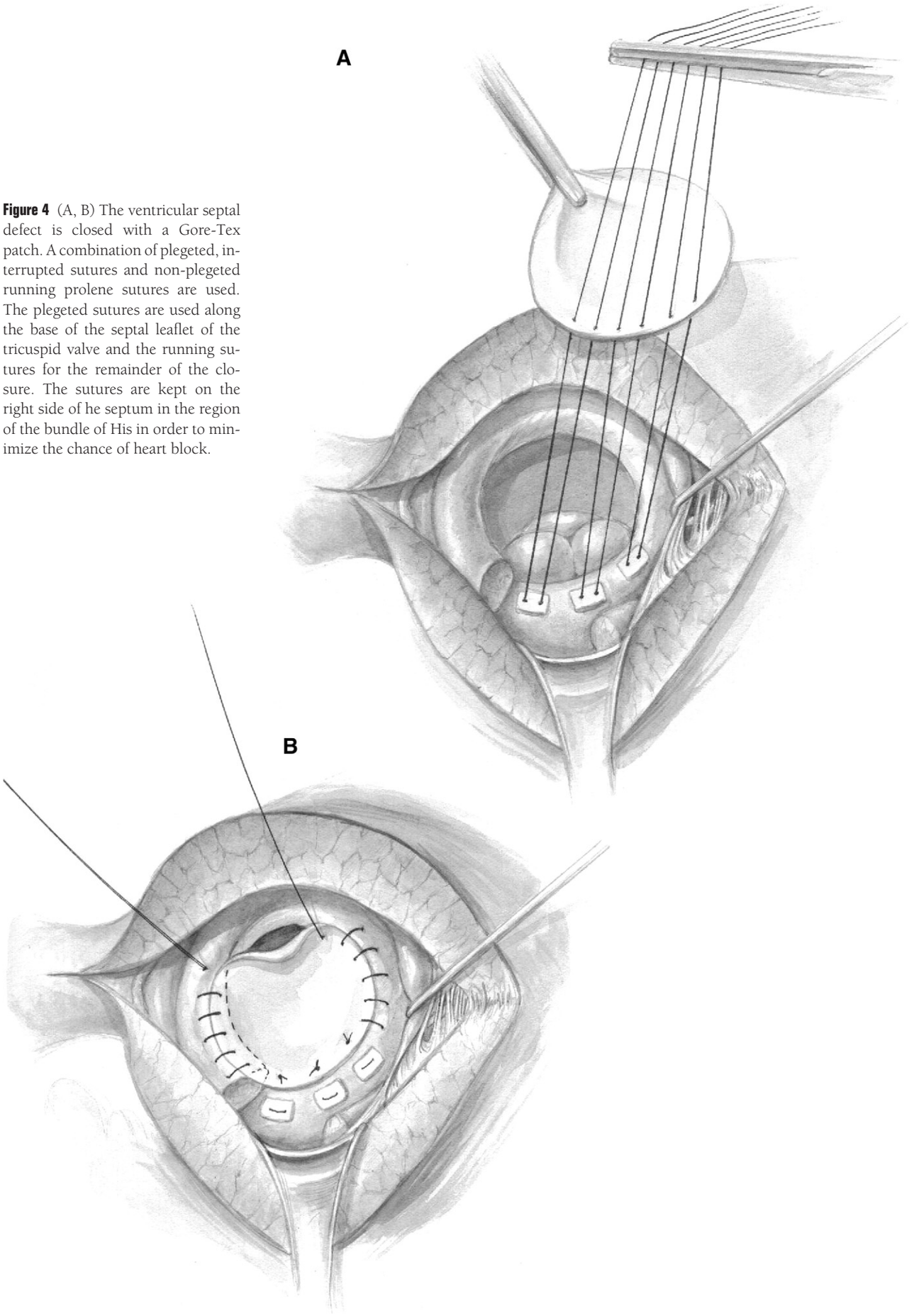

Figure 4 (A, B) The ventricular septal defect is closed with a Gore-Tex patch. A combination of plegeted, interrupted sutures and non-plegeted running prolene sutures are used. The plegeted sutures are used along the base of the septal leaflet of the tricuspid valve and the running sutures for the remainder of the closure. The sutures are kept on the right side of he septum in the region of the bundle of His in order to minimize the chance of heart block.

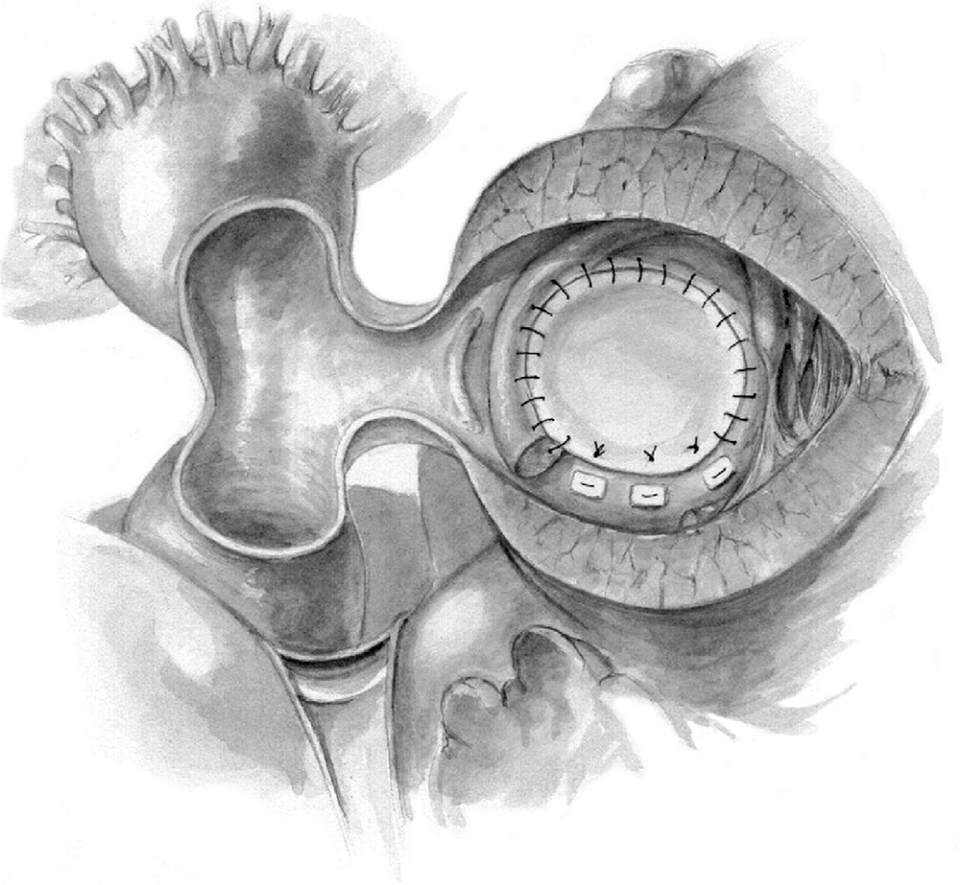

Figure 5 The main pulmonary artery incision is carried distally to expose the pulmonary artery bifurcation.

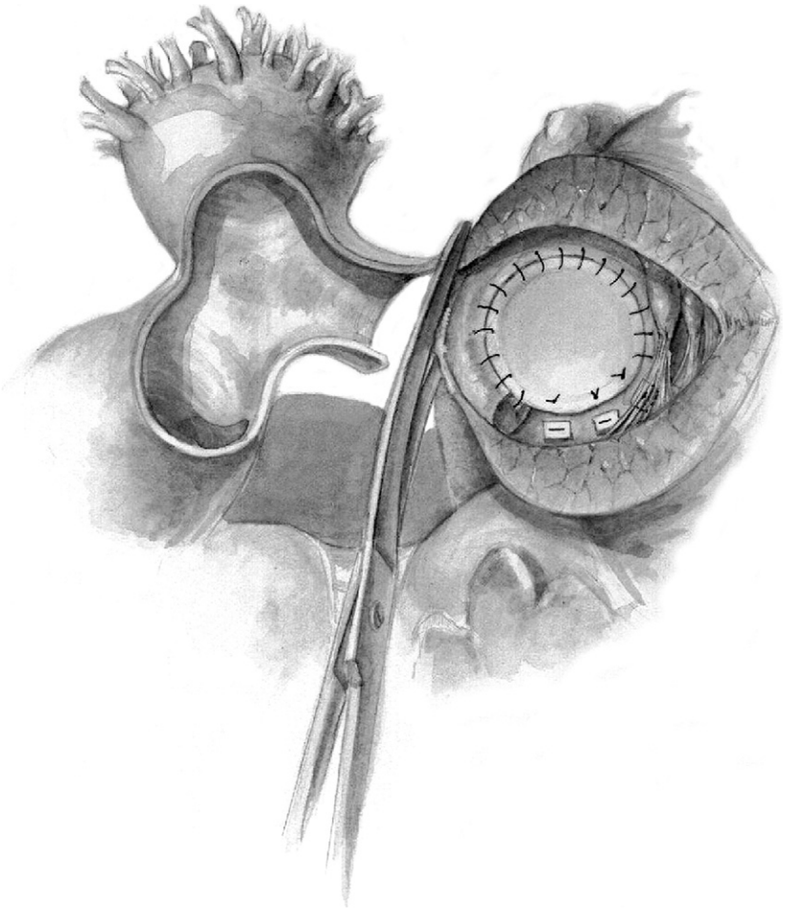

Figure 6 The main pulmonary artery is detached from the right ventricle at the level of the pulmonary valve annulus.

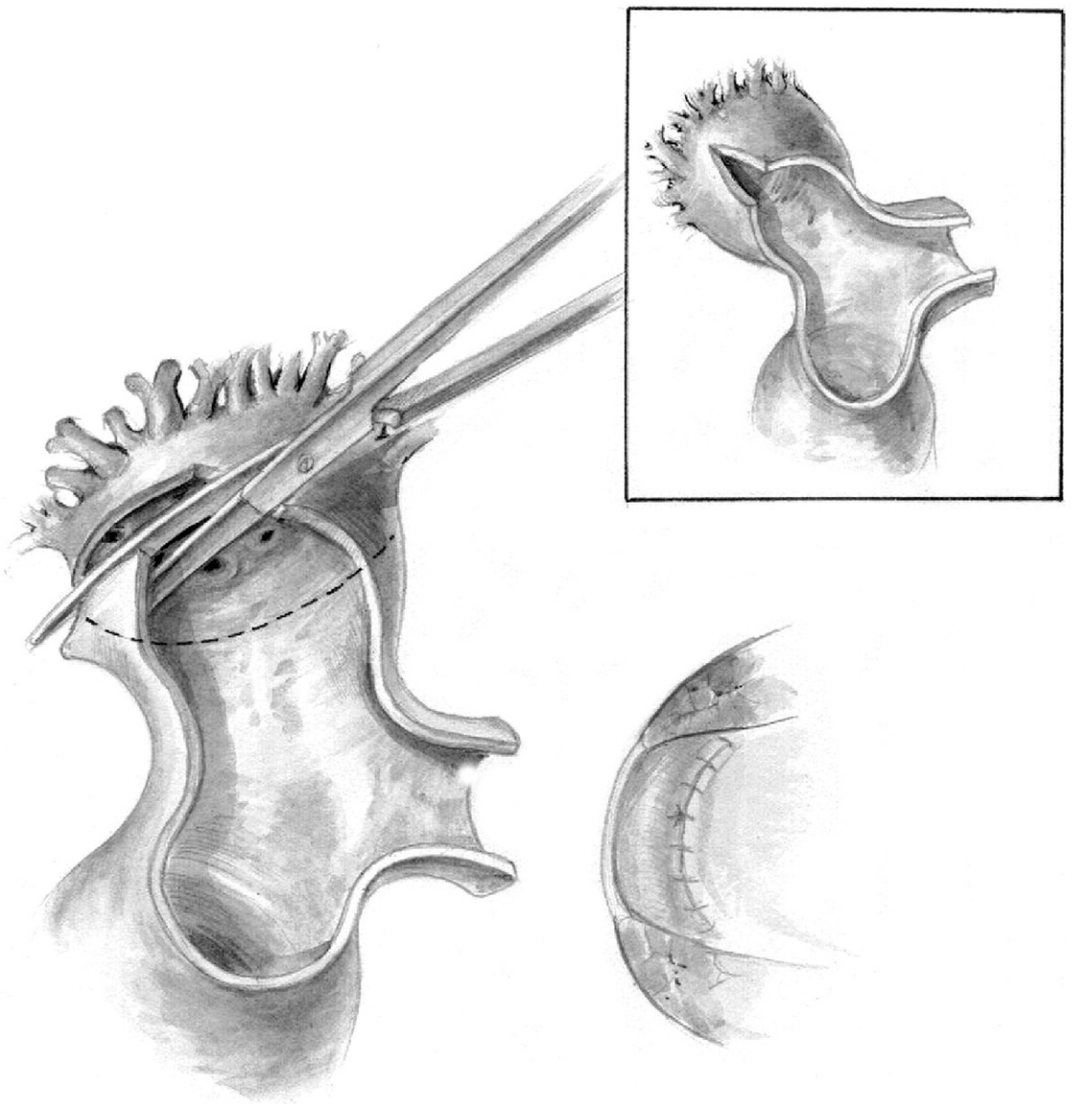

Figure 7 The left pulmonary artery is divided leaving only a narrow cuff of tissue at the level of the origin of the lobar branches.

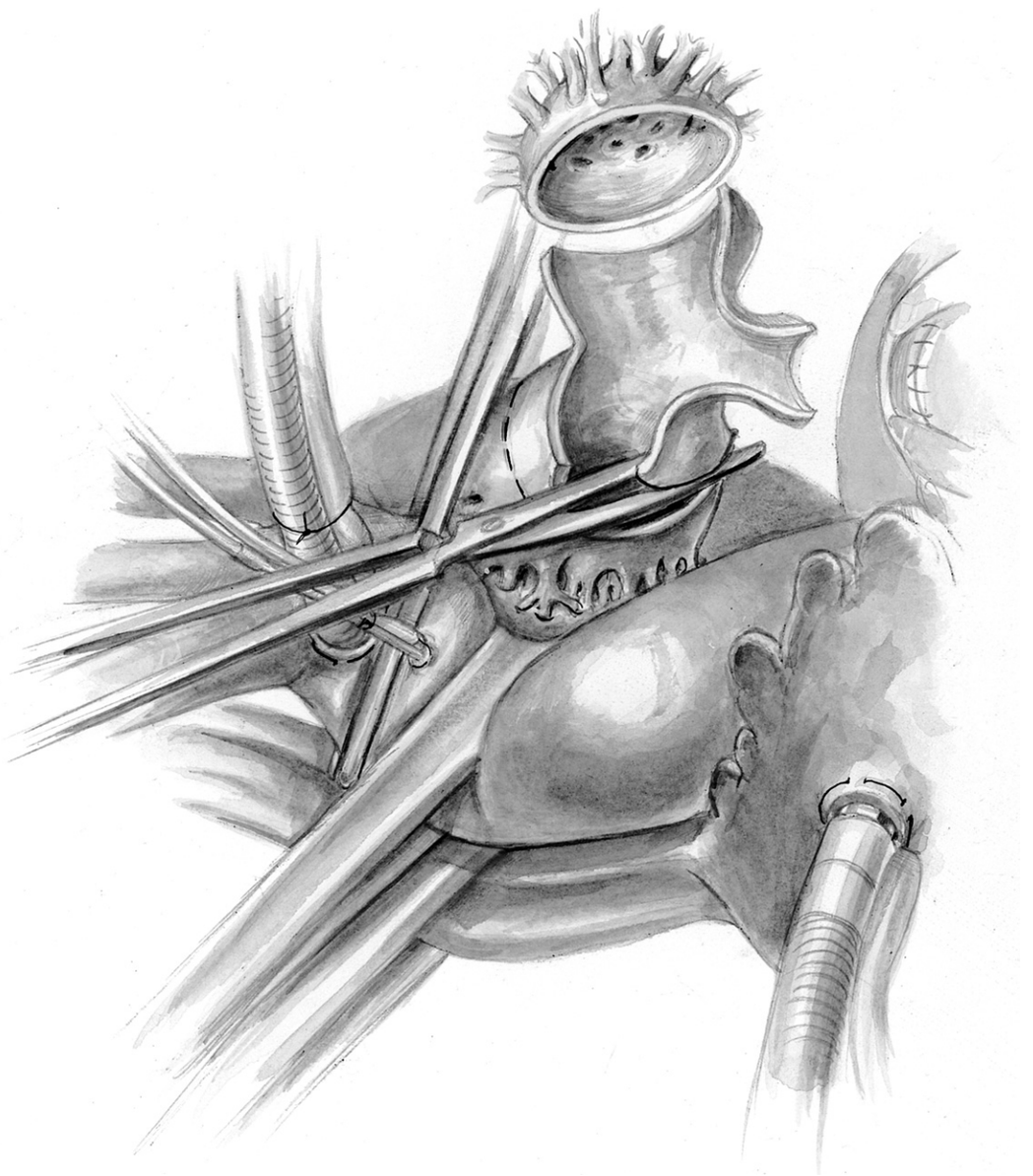

Figure 8 The right pulmonary artery is divided at the same level resulting in the removal of almost all abnormal pulmonary artery tissue.

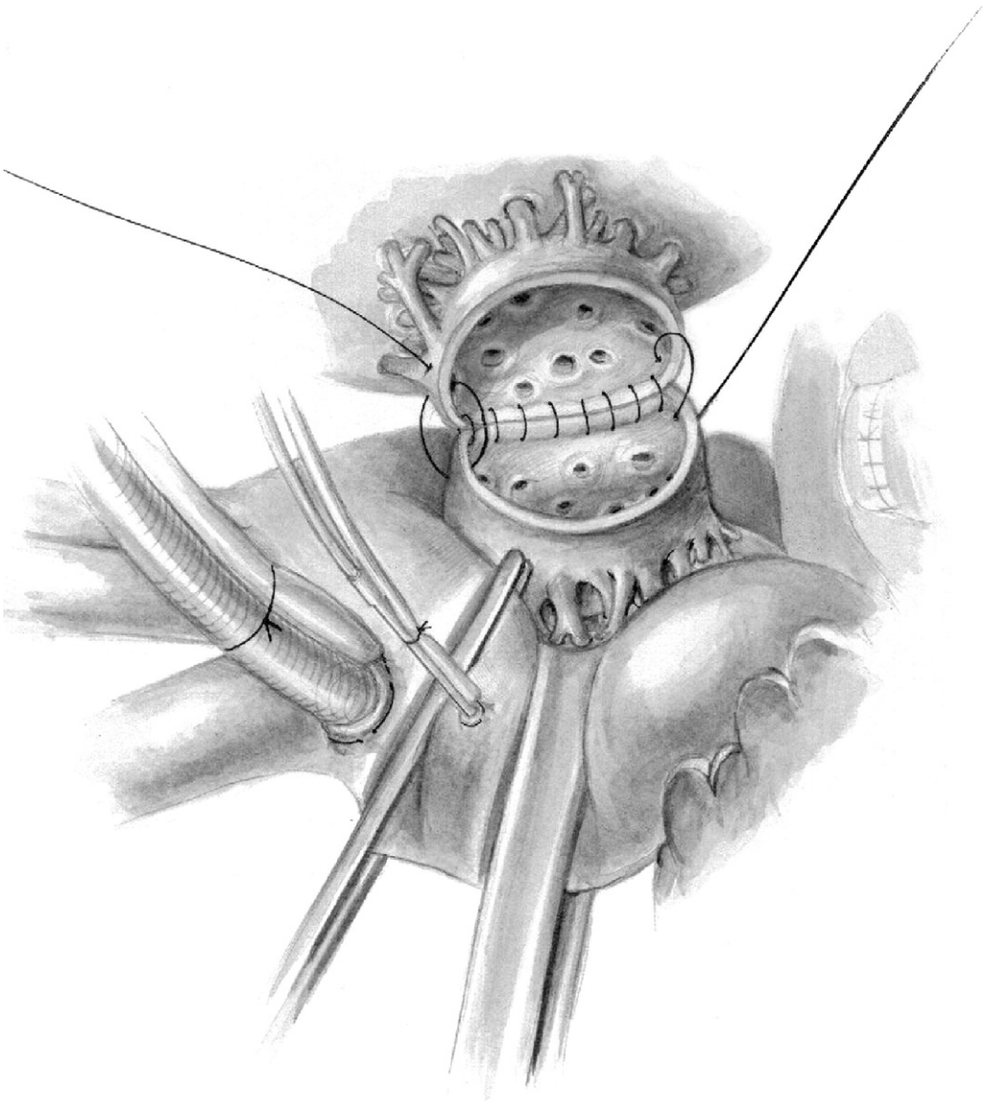

Figure 9 The posterior walls of the divided right and left pulmonary arteries are then approximated with a running prolene suture. Mobilization of the branch pulmonary arteries facilitates this anastamosis. The relative short distance between the divided branch pulmonary arteries allows for a tension free anastamosis in the neonate. In older patients, we occasionally leave more posterior wall tissue in order to facilitate a tension free anastomosis.

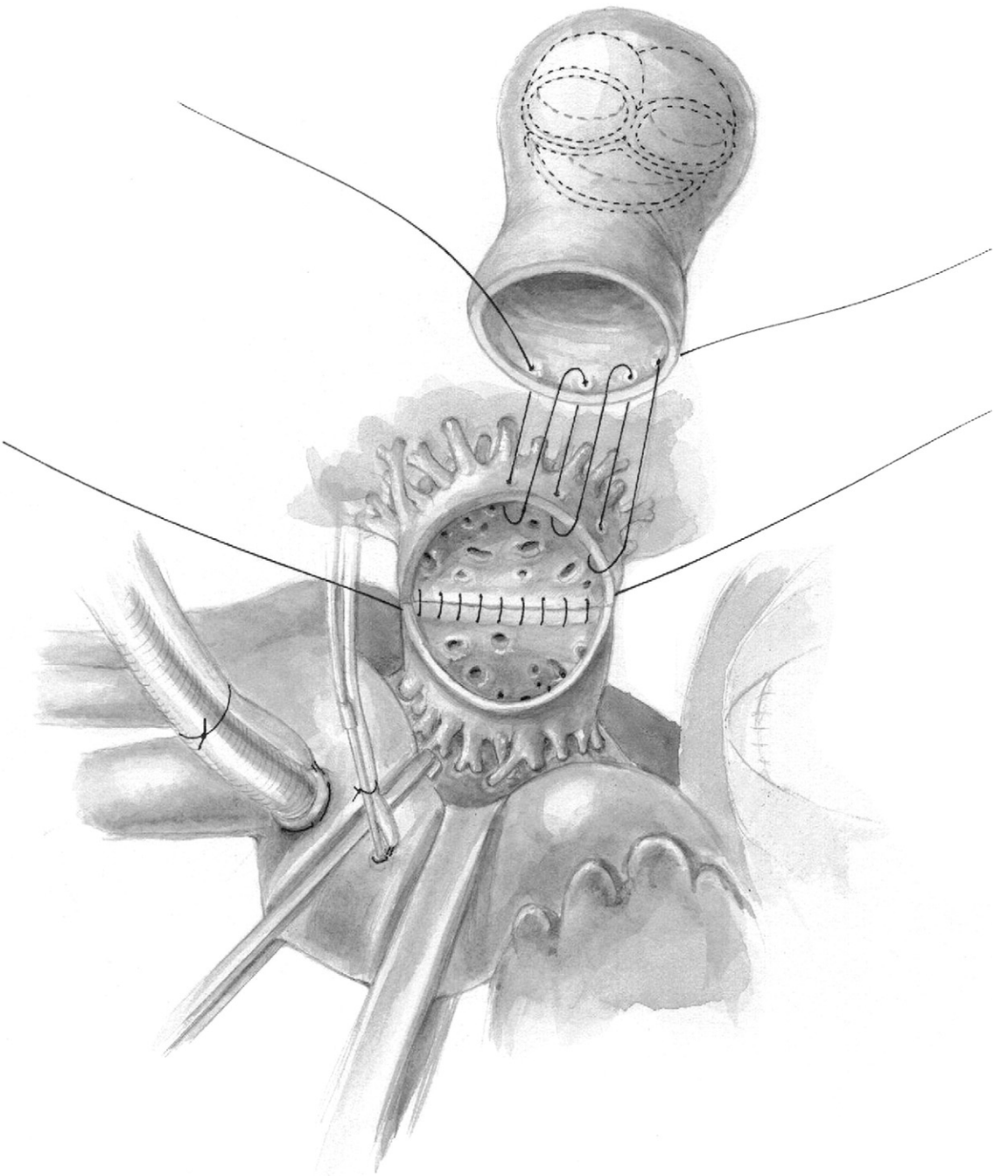

Figure 10 An appropriate sized pulmonary homograft is anatamosed to the pulmonary artery reconstruction distally using a running prolene suture. During this time, the patient is rewarmed and at the completion of the suture line, the left side of the heart is deaired and the aortic cross clamp is removed.

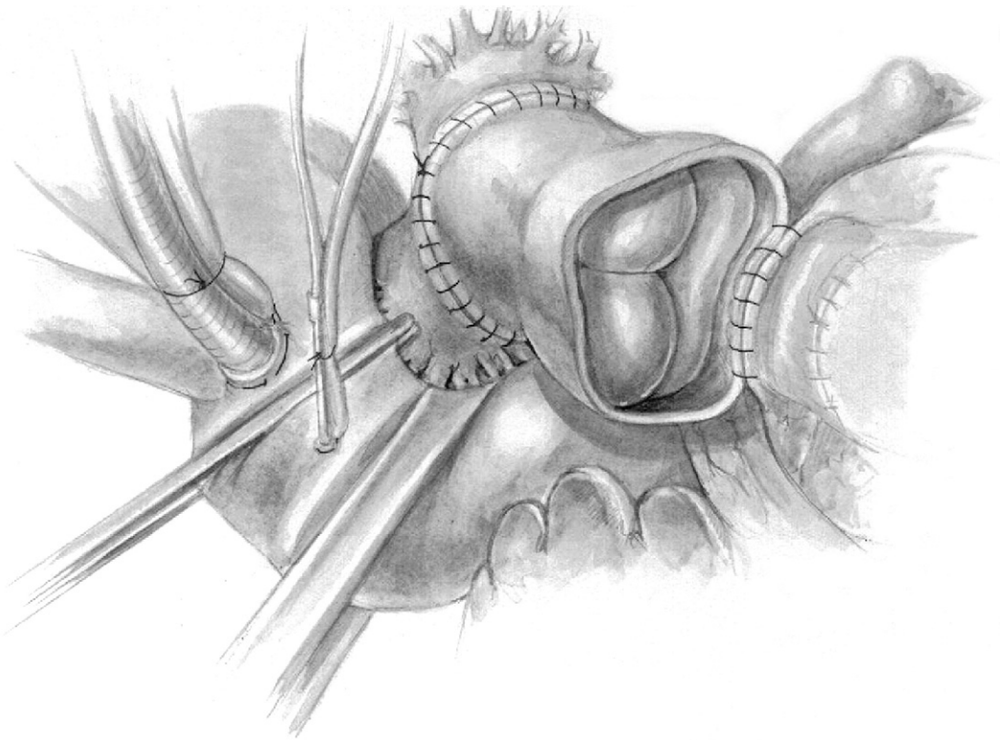

Figure 11 The proximal end of the homograft is anastomosed to the infundibulum of the right ventricle.

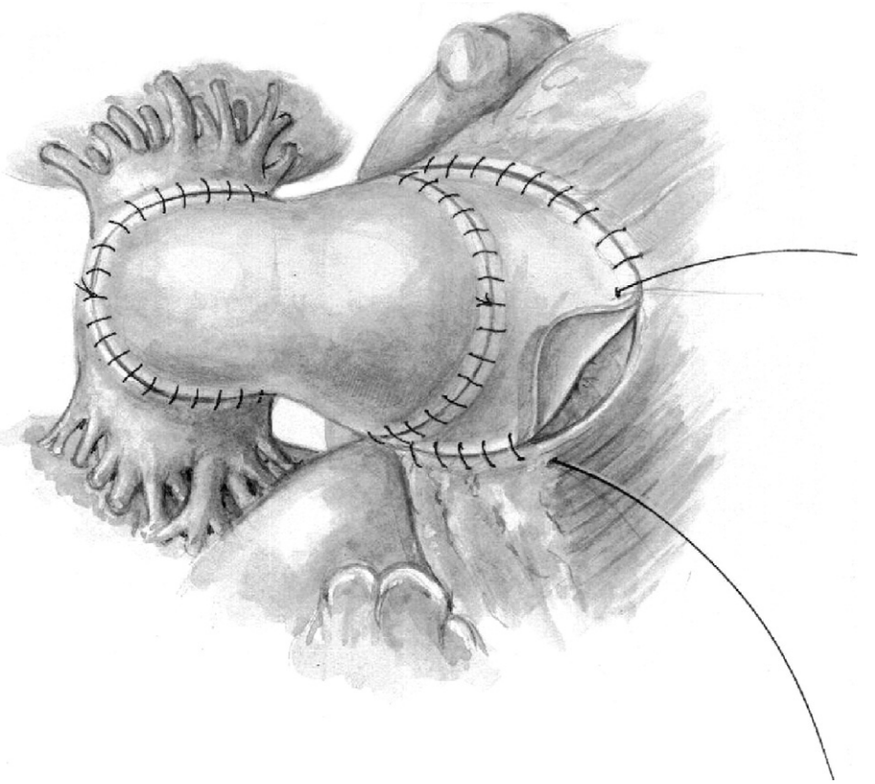

Figure 12 The pericardium that was harvested at the beginning of the operation and placed in glutaraldehyde is now used to augment the right ventricular outflow tract so there is no obstruction. The right side of the heart is deaired prior to completion of this suture line. The foramen ovale is left open in order to allow right to left decompression in the immediate postoperative period. If a secundum atrial septal defect is present we downsize it to 4 millimeters by cinching a purse string suture around an appropriately sized dilator. A left atrial line is left in place for monitoring. Two atrial and two ventricular pacing wires are placed as well. The patient is weaned from cardiopulmonary bypass and the adequacy of the repair is checked with transesophageal echocardiography.

Conclusions

The repair described above accomplishes two principles that we feel contribute to better patient outcomes: excision of nearly all aneurysmal pulmonary artery tissue and placement of a valve in the pulmonary outflow tract. Resection of almost all aneurysmal tissue minimizes both the risk of bronchial compression and the chance of future aneurysmal dilation. Placement of a valve in the pulmonary outflow tract protects the right ventricle from the negative effects of free pulmonary insufficiency and simplifies the management of postoperative pulmonary hypertension. The use of circulatory arrest with intermittent perfusion provides the benefits of circulatory arrest without prolonged periods of cerebral ischemia. To further reduce the risk of adverse neurological outcomes, we have begun to base the length of arrest periods on near infrared spectroscopy measurements of cerebral oxygenation rather than using arbitrary time periods. This technique for repair of TOF-APVS is relatively simple and reproducible and causes minimal distortion of the pulmonary arteries.

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